Maryland
DEPARTMENT OF HEALTH

Ravicti (glycerol phenylbutyrate)

Ravicti is a nitrogen-binding agent indicated for chronic management of patients with urea cycle disorders (UCDs) who cannot be managed by dietary protein restriction and/or amino acid supplementation alone.

Limitations of Use:

- Ravicti is not indicated for treatment of acute hyperammonemia in patients with UCDs.
- Safety and efficacy for treatment of N-acetylglutamate synthase (NAGS) deficiency has not been established.

I. Criteria for Initial Approval

Ravicti will be considered for coverage when **ALL** of the criteria below are met, confirmed with supporting medical documentation.

- Approved for both pediatric and adult patients.
- Ravicti should be prescribed by a physician experienced in management of UCDs or in consultation with, a board-certified geneticist/metabolic specialist or physician experienced in the management of urea cycle disorder.
    - Prescribed with active involvement of a nutritionist to maximize caloric intake with neutral nitrogen balance.
- Documented confirmed diagnosis of a urea cycle disorder (UCD).
    - As evidenced by plasma ammonia concentration of 150 micromole/L or higher associated with a normal anion gap and a normal plasma glucose concentration and molecular genetic testing or measurement of enzyme activity.
    - Patient has documented history of hyperammonemia associated with diagnosis of a UCD as one of the following:
        - Carbamoyl phosphate synthetase 1 deficiency (CPS1D);
        - Orthinine transcarbamylase deficiency (OTCD);

        - Argininosuccinate synthetase deficiency (ASSD/classic citrullinemia/type 1 citrullinemia);
        - Argininosuccinate lyase deficiency (ASLD/argininosuccinic aciduria); OR
        - Arginase deficiency (ARG1D/argininemia).

- Patients must have tried and failed Buphenyl (sodium phenylbutyrate).

    - Patient has a comorbid condition that prohibits a trial of Buphenyl (sodium phenylbutyrate) due to its sodium content.
    - Patient can not tolerate Buphenyl due to severe adverse effects.
    - Patient has a contraindication to Buphenyl (e.g. hypersensitivity, pregnancy, breastfeeding.)

- Patient's condition cannot be managed by dietary protein restriction and/or amino acid supplementation alone.
    - Raviciti must be used with dietary protein restriction and, in some cases, dietary supplements.
- Patient must currently be treated with, and adherent to dietary protein restriction, and when appropriate, dietary supplements (e.g. essential amino acids, arginine, citrulline, protein-free calorie supplements.)

- Documentation of addressing any possible complications and need for medication discontinuation, associated with concurrent drug use when appropriate.

    - Corticosteroids, valproic acid (Depakene, and others), and haloperidol (Haldol, and generics): May increase plasma concentrations of ammonia.

    - Probenecid: Concomitant use of probenecid may decrease excretion of phenylacetate and phenylacetylglutamine.

II. Criteria for Continuation of Therapy

All of the criteria for initial therapy (**in Section I.**) must be met; **AND** the provider attests to a positive clinical response.

III. Dosing/Administration

Ravicti must be administered according to the current FDA labeling guidelines for dosage and timing.

IV. Length of Authorization for Initial Therapy

Ravicti will be authorized for 12 months when criteria for initial approval are met. Continuing therapy with Ravicti will be authorized for 12 months.

V. Billing Code/Information

J8499 Prescription drug, oral, non chemotherapeutic, not otherwise specified.

Prior authorization of benefits is not the practice of medicine nor the substitute for the independent medical judgment of a treating medical provider. The materials provided are a component used to assist in making coverage decisions and administering benefits. Prior authorization does not constitute a contract or guarantee regarding member eligibility or payment. Prior authorization criteria are established based on a collaborative effort using input from the current medical literature and based on evidence available at the time.

Approved by MDH Clinical Criteria Committee: 1/27/2021
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